Cochlear Implants in Children

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INTRODUCTION

In recent years, notable progress has been made in the rehabilitation of children with mild to moderate-severe hearing impairment using new and advanced hearing aids. The major problem which remained was the lack of availability of treatment options for those with severe-to-profound sensorineural hearing loss who received little or no benefit from conventional amplification. The purpose of the cochlear implant is to provide these children with direct electrical stimulation of the auditory nerve. A hearing aid amplifies incoming stimuli while a cochlear implant attempts to replace a function lost by the cochlea. In a normal hearing ear, the hair cells within the cochlea act as a transducer of mechanical energy of sound vibration to energy capable of enervating the eighth nerve. The consequence of a decrease in the number of hair cells is the loss of ability of the cochlea to perform the functions, which result in eighth nerve stimulation. The implant replaces the task of the lost hair cells by converting mechanical energy into the electrical energy necessary to excite the remaining cochlear neurons.

In order to truly appreciate the application and unfolding of cochlear implants as applied to the pediatric population, it is important to summarize the development of the devices in general.

DESIGN EVOLUTION

The concept of electrically stimulating the auditory nerve was first explored by Volta around 1800. In the 1950's, Djourno and Eyries reported on the first stimulation of the eighth nerve in an adult; there was substantial skepticism and criticism, however, regarding the safety and efficacy of inserting an electrical device into the cochlea. In the 1960's, Dr. William House capitalized on new surgical techniques as well as the reports of Djourno and Eyries, to experiment with direct stimulation of the cochlea. The rapidity of development of cochlear implant devices for clinical purposes was made possible, in part, by the adaptation of the technology used in pacemakers including biocompatible materials and electrode design.

Although there have been many variations on the theme, the basic design of an implant system has remained relatively stable over the years. It consists of an external microphone, processor and transmitter and an internal receiver-stimulator and electrode array. The microphone captures incoming sound and converts it into electrical signals. The processor configures, amplifies and manipulates the electrical signal into the preferred paradigm, which is then transmitted to the internal receiver/stimulator and electrode array. Subsequently, the electrodes are stimulated in a pattern, which is determined by the encoding strategy of a given prosthesis.

In the approximately 35 years of cochlear implant development, various configurations and encoding strategies have materialized. From a design perspective, cochlear prostheses have evolved from extra-cochlear (the electrode array does not invade the cochlear space), single-channel single-electrode systems to intracochlear (electrode array is placed in the cochlea) multi-channel multiple electrode arrays. The transmitter can be percutaneous or transcutaneous: A percutaneous transmitter connects directly to the internal receiver through an electro-mechanical connection while a transcutaneous system delivers the signal via radio frequency (RF) linkage. The progression of coding strategies has been more variable. In all implants, the incoming stimulus is converted into an electrical signal. Early single-channel, single-electrode systems did not require extensive shaping or coding of incoming sound: the stimu-

lus was simply converted into an electrical signal and the sole electrode was stimulated. An example of an analog driven system is the Simultaneous Analog System (SAS) available in the Clarion device (Figure 1) where analog signals are transmitted concurrently to individual electrodes or electrode pairs in a bipolar mode (current flows between an active electrode and a ground electrode). Other factors involved in the determination of the pattern of electrode excitation are simultaneous (several electrodes can be stimulated at the same time) versus sequential stimulation (electrodes are stimulated sequentially) and waveform representation (filter bank) strategy versus the 'n of m' strategy. In the 'n of m' strategy, where 'n' refers to the number of channels and 'm' refers to the channels with the greatest amount of energy. In this strategy, the incoming signal is divided into the six to ten channels that have the greatest amount of energy and the appropriate electrodes are then stimulated. The signal is scanned every few milliseconds for these peaks to effect the greatest possible fidelity. Currently, versions of this processing mode are implemented in the Nucleus 22 and CI24M devices (SPEAK) (Figure 2) and the MedEl cochlear implant (Figure 3). The CIS (continuous interleaved sampling) encoding strategy is a nonsimultaneous pulsatile system where the stimuli are interleaved though non-overlapping and transmitted rapidly and sequentially to decrease electrode interac-

Figure 1. Clarion S-series speech processor and implantable cochlear stimulator (ICS).

tion while increasing data transmission. It employs a filter bank/waveform representation of the speech stimulus, as opposed to the speech feature model employed in implementations of the 'n of m' coding system.

Electrode design has been another area of development and difference between devices over the years. The Nucleus and MedEl internal arrays have traditionally employed a straight free-fitting electrode design whereas the Clarion array is coil-shaped, intended to hug the modiolus. The concept behind modiolar hugging electrode designs is placement closer to the intended stimulation site in order to reduce the amount of power necessary to effect neural stimulation. A study by Roland et al (in press), however, revealed that no currently commercially available electrode design is, in fact, modiolar-hugging although research is presently being conducted to develop electrode arrays, which would stimulate the neurons more effectively. At this point, however, the precise impact of the various electrode designs and stimulation modes on patient performance is still unknown.

COCHLEAR IMPLANTS IN ADULTS

No discussion of cochlear implants in children would be complete without laying the foundation with its forerunner: cochlear implants in adults. Since the focus of this chapter is children, the review will be necessarily brief but the reader is referred to several published chapters by Luxford and Brackmann (1985) and House and Berliner (1991) which offer detailed historical background.

The first human clinical implantations in the United States began in the 1970's with devices developed by Michelson (1971) and House and Urban (1973). These investigators, plus Simmons at Stanford University, were instrumental in advancing the cause of cochlear implantation in the face of much criticism and skepticism. Most of the focus in the early to middle 1980's was to establish the efficacy and safety of both single- and multichannel devices in profoundly deaf adults. This accomplished, comparisons of systems began in order to determine the assumed supremacy of multi-channel, multi-electrode prostheses. Reports by Gantz et al (1988) and Waltzman et al (1992) confirmed that multi-channel devices enabled postlingually deafened adults to obtain significantly more open-set word and sentence recognition than was possible with single-channel devices. Over the years, several multi-channel systems have been commercially available; the early models included
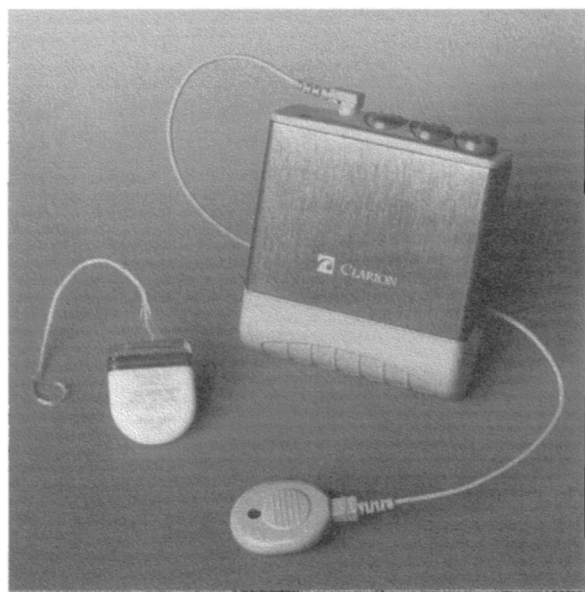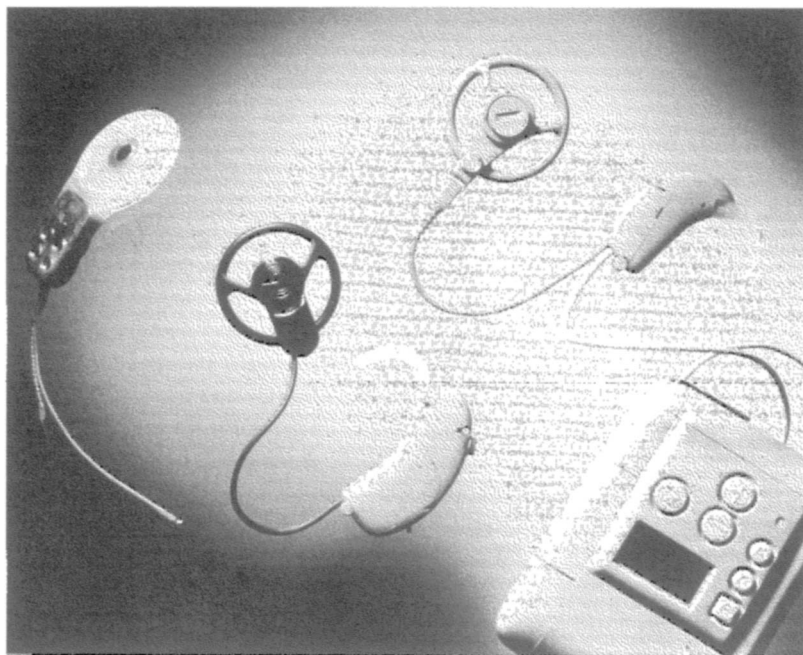
Figure 2. Nucleus CI24M ear-level and body-worn speech processor, and receiver-stimulator and electrode array.

the Ineraid, Storz and Nucleus (F0F1F2). The Ineraid and Storz devices are no longer manufactured and the Nucleus device has been upgraded several times and currently uses technologically advanced processing schemes. In addition to the Nucleus implant, the Clarion and MedEl multichannel prostheses are currently available for use in the United States: additional multichannel devices, including the French manufactured Digisonic, are used outside of the United States. Concurrently, as the signal processing capabilities have advanced, the criteria for implantation of adults has expanded to include those with severe to profound hearing losses and some open-set speech recognition and the congenitally and prelingually deafened. Adult implantees have gone from expecting primarily an aid to lipreading and obtaining only minimal amounts of word and sentence open-set speech recognition post-implantation to achieving open-set discrimination scores between 70%–100% on all measures, enabling many of them to communicate more efficiently, use the
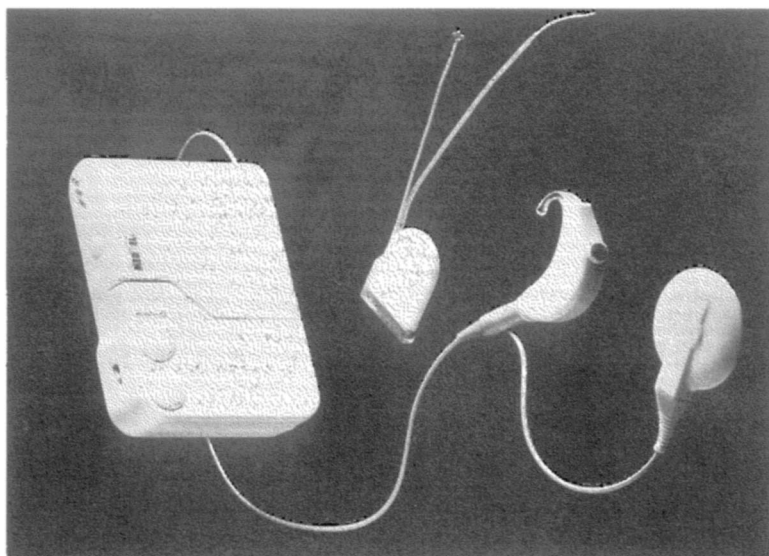
Figure 3. MedEl Combi 40+ cochlear implant and CISPRO speech processor.

telephone and resume professional activities. The evolution of cochlear implants over the past thirty years has undoubtedly dramatically changed the quality of life of adult recipients.

During the 1980's, as the results in adults improved with each successive technological improvement, a serious interest in children began to emerge. Although single-channel devices were being implanted in children as part of a clinical trial, the postoperative performance was not commensurate with the outcomes being seen with adults using multi-channel implants. It was evident that a concerted effort was needed to explore the possibility of expansion into the pediatric population. In February 1986, a conference was held in Colorado to examine and discuss the various issues involved with pediatric cochlear implantation. Although in 1999 it seems naive to discuss patient selection, evaluation techniques, safety issues and other pertinent areas, thirteen years ago these topics were controversial in relation to the application of cochlear implants as a treatment for deafness in children. Participants in the symposium were divided into committees which discussed and recounted to the group a variety of topics related to implantation of children. The colloquium attempted to put forth selection criteria which might relate to successful implantation in children including length of deafness and age at implantation. Surprisingly, considering the current acceptable younger boundary for age of implantation, the group felt that accurate auditory thresholds could not be obtained on very young children and, therefore, suggested a lower age limit of two years for implantation. As the reader will see further on in this review, the opinion on this has shifted considerably. Further, the group considered psycho-social issues, educational environment, rehabilitation strategies, speech perception and language, and speech production assessments as vital to the process. The need to measure improved speech perception over time was emphasized as crucial in order to validate the use of implants in children and to correlate improved auditory perception with language and speech development. The surgeon's group recommended surgical training for each separate type of electrode array and believed that, from a surgical standpoint, two-year-old children were acceptable candidates for implantation. In summary, this colloquium shared thoughts and concerns which formed a basis for current concepts related to the implantation of children.

By the mid-1980's, two cochlear implants were undergoing FDA approved clinical trials for use in children in the US: the 3M/House single-channel implant and the Nucleus multi-channel 22-electrode device. The 3M/House device used monopolar stimulation (ground electrode is outside the cochlea) and consisted of a single platinum ball electrode which was inserted approximately 6 mm into the scala tympani with the titanium receiver acting as the ground and a 340 to 2700 Hz filtered signal amplitude modulated with a 16 kHz inaudible carrier. The decision to proceed with implantation in children was based on the relatively few complications found in adult implantees and the belief that the increased access to sound provided by the implant could assist the child in speech and language development. Candidates were restricted to those children with documented profound hearing loss and prior hearing aid experience. The preoperative and postoperative assessment tools, in addition to routine sound field audiometric measures, were closed set tests which focused primarily on the suprasegmental aspects of speech signal but included some speech comprehension tasks. Indeed, it was quite different from the current emphasis on open-set phoneme, word and sentence recognition. Postoperatively, the majority of children had increased access to sound in general and environmental sounds, in particular. Scores on closed set identification tests improved with time and a few children who were tested on simple open-set measures after several years of usage showed some ability to repeat simple phrases without the benefit of lipreading. Unfortunately, the lack of consistent and rigorous subject and data collection and analysis methods impacted negatively on the claims that could be made regarding the results with the 3M/House device. Interestingly, however, the factors that were associated with better postoperative performance in the 3M/House pediatric implantees are variables which currently affect open-set speech perception with multi-channel prostheses including length of deafness, age of onset, mode of communication and length of device usage: The longer the device usage, the shorter the length of deafness, the earlier the onset of deafness, the use of oral communication have all been associated with better outcome. More on factors affecting performance later.

Concurrently, in 1987, the Nucleus 22 multichannel multi-electrode device became available for use in children under a FDA sponsored clinical trial. The rationale for allowing its use in children was the proven efficacy and safety in adults.

The processing strategy implemented in the early devices transmitted fundamental frequency and second formant information with first formant information added shortly thereafter. The basic premise was that there are certain features of the speech signal which when identified would provide the necessary information to identify the entire stimulus-hence the term 'speech feature' encoding strategy. As with the House/3M device, candidates were restricted to those with bilateral profound sensorineural hearing loss. Some of the study design flaws identified with the study of the 3M/House device were rectified with the protocol for the Nucleus device permitting more identifiable and justifiable conclusions. The transmission of the additional timing and intensity information afforded by the multi-channel system resulted in rapid progress and more consistent development of open-set auditory skills in pediatric implantees. Initially, data collection centered around establishing safety and limited efficacy with 'limited' simply referring to the very cautious optimism that pervaded the implant community. Expectations revolved around improved access to sound, aid to lipreading and improved closed-set word identification. Within a short period of time, however, it became clear that attainable goals were far above the very conservative expectations. At this point, assessment tools which measured open-set speech recognition became central to the evaluation process.

Any discussion of cochlear implants in children must be divided into several central areas: criteria for implantation, assessment, surgery, device programming, speech production, language development, mode of communication, education and outcome measures related to the processing strategy used in a given prosthesis. Since these, and additional variables, interact to affect the postoperative performance in children, this monograph will attempt to review the development and current status of each of the factors and how they correlate with one another.

CRITERIA FOR IMPLANTATION

Table 1 outlines the current criteria for implantation of children using FDA approved devices. The protocols for earliest FDA sponsored clinical trials using the 3M/House and Nucleus 22 devices required that prospective candidates 1) be at least two years of age, 2) have a bilateral profound sensorineural hearing loss, 3) obtain no measurable benefit from appropriate amplification, and 4)

Table 1. Criteria for pediatric implantation of FDA approved devices

1. Clarion Multistrategy Cochlear Implant (1997)
— 2–17 years of age. Children as young as 18 months may be implanted with cause.
— Bilateral profound sensorineural hearing loss.
— Trial period with hearing aids.
— Lack of significant gains from amplification as defined by a score of 0% on the Phonetically Balanced Kindergarten (PBK) word test. In younger children, a lack of benefit is defined by the lack of consistent response to auditory stimuli as measured on the Meaningful Auditory Integration Scale (MAIS).

2. Nucleus 22 Multichannel Cochlear Implant (1990)
— 2–17 years of age.
— Bilateral profound sensorineural hearing loss.
— Little or no measurable benefit from amplification.

3. Nucleus CI24M (1998)
— 18 months–17 years of age.
— Bilateral profound sensorineural hearing loss as defined by little of no benefit from appropriate amplification. The MAIS and Early Speech Perception Test (ESP) can be used to document the lack of development of auditory skills in young children. In older children, the lack of benefit is defined as a score of 20% or less on the Lexical Neighborhood Test (LNT) test. All children are required to undergo a trial period with hearing aids.

have no psychological or medical contraindications to implantation. The parents, and the child if he/she was older, needed to have realistic expectations as to the possible postoperative benefits of implantation. For example, if the parents and/or child expected that telephone usage was either a foregone conclusion or a necessary precursor to implantation, then counseling was in order prior to proceeding with the process. A medical contraindication might include an individual who for some reason could not have general anesthesia. Additionally, the parents had to be willing to return to the implant center to fulfill the protocol requirements. When the Clarion device was approved for clinical trials in children, the criteria for implantation were essentially the same as those for the Nucleus 22: minimal functional benefit with hearing aids was defined as a score of 0% on the Phonetically Balanced Kindergarten (PBK) monosyllabic word test. By the time the Nucleus CI24M became available to children under an FDA clinical protocol in 1996, the inclusion criteria were far less restrictive: the lower age limit became 18 months and the monosyllabic word score as measured by the Lexical Neighborhood Test (LNT)

had to be less than 20% as compared to 0% on the PBK test. The relaxing of the criteria can be attributed to the impressive results seen over time in pediatric implantees who were previously implanted with the Clarion or Nucleus 22 devices. In truth, children as young as 12 months old have been implanted in the United States. Initially, decisions to proceed with surgery at these very young ages had been associated with post-meningitis labyrinthitis ossificans where the ability to place the electrode array in the proper position could be jeopardized by bony growth. Currently, some congenitally deaf 12-month-olds are being implanted despite the absence of a progressive disease process: the 'earlier is better' theory has clearly evolved although documentation of improved performance on these very young implantees is not yet available.

Lowering the age criteria for implantation is one issue: the second expansion issue involves the implantation of children who have more functional residual hearing. Gantz et al (1998) reported on the results of a group of profoundly hearing impaired children (mean PTA = 108 dBHL) with preoperative word recognition scores of 4–24% in the best aided condition. At the two year postoperative interval, these children had a mean percent correct score of 75% implant alone equating their performance with a group of moderately hearing impaired hearing aid users with a PTA of 71 dBHL.

It appears that the natural evolution for expanding the criteria for implantation should include systematic studies and trials of the long-term benefits and effects of implanting both very young children and those with functional residual hearing. There is every reason to believe that the trend towards wider criteria windows will continue.

ASSESSMENT

The concept exists that appropriate and accurate assessment of the degree and type of hearing loss, and accompanying effects on speech perception production and language development, underlies our ability to use cochlear prostheses as an intervention tool in the pediatric population. This notion is even more critical with the possibility of implantation developing into a reality for children under the age of 18 months and as young as 10 months.

Individual ear thresholds are integral to the diagnostic protocol. The preferred measurement method is behavioral since it does not require invasive techniques and provides the best indication of the scope of pure-tone hearing. Although it is commonly thought that obtaining accurate individual ear thresholds is impossible in infants, experienced audiologists have been able to obtain relatively accurate and complete audiograms in infants as young as 4 months of age; however, in many situations involving either the very young or multiply-handicapped child, objective measurement techniques need to be used in order to obtain valid thresholds. In these more difficult cases, frequency-specific auditory evoked potentials (AEP) are often employed and behavioral measures are used to confirm the results when the child is able to perform the task. Since current AEP techniques have evolved to a degree which allows definitive diagnosis to occur, a child can be diagnosed with a hearing loss as a neonate allowing for appropriate intervention to commence immediately. The early diagnosis coupled with a trial period of appropriate amplification increases the potential for implantation at an early age. The measurement of auditory thresholds, however, is only the beginning of the task.

As we are all aware, pure tone thresholds do not necessarily reflect functional ability. Some profoundly deaf children have excellent speech perception abilities using hearing aids while others with seemingly identical 'hearing' have no ability to understand words or sentences in an auditory-only environment. Since the primary purpose of the cochlear implant is to enhance the ability of the deaf to understand speech, it is necessary to have tools which can accurately measure speech perception skills. Although non-speech based material is often used to study and assess the fundamental concepts of perception in adults, these procedures are generally too complex and advanced for children: assessment of speech perception in children is best accomplished by employing age appropriate speech stimuli in a variety of contexts since one test would be unlikely to be capable of assessing all aspects of speech reception and perception in children of all ages.

The evaluation of speech perception can be divided into several levels: suprasegmental and segmental aspects of the speech stimulus, word or sentence materials and closed- or open-set response formats. In the early days of cochlear implantation of children, the evaluation centered on the suprasegmental and closed-set formats. The caution was related to the lack of knowledge regarding the nature and limits of postoperative performance in children: were the prognostic indicators

and expectations associated with adults valid for the pediatric population as well? Since the only cochlear implant available to children in the United States in the early-to-mid 1980's was the single channel 3M/House device, the assessment tools were primarily based on the observations and expectations with that particular prosthesis. The two most widely used tests, which focused mainly on the stress and intonation patterns of the speech signal, were the Discrimination After Training (DAT) and the Monosyllable-Trochee-Spondee test (MTS). The DAT was developed in 1984 by Thielemeir and consists of twelve hierarchical levels which measured the detection of a speech sound, timing, duration and stress patterns and word identification using a closed-set format. The MTS (Erber and Alencewicz, 1976) assesses the ability of the child to correctly identify syllabic stress patterns and words. The stimuli are twelve words of varying syllables: the child is required to identify the member family of a given stimulus (monosyllable, two-syllable word with equal energy and two-syllable word with unequal energy). The percent of words categorized correctly is the stress score and the number of words identified correctly from a choice of four words is the word score.

Several additional tests, which although not specifically designed for the purpose, include subtests to assess suprasegmental competence. The Test of Auditory Comprehension (TAC) evaluates various aspects of perception using both non-speech and speech stimuli (Trammell et al, 1981). Subtests 1 to 3 tests noise versus voice, human versus environmental differentiations and stress, rhythmic and intonation competence. The Glendonald Auditory Screening Procedure (GASP) and the Early Speech Perception test (ESP) also have subtests dedicated to assessing suprasegmental characteristics of the speech signal but were more widely used for word identification tasks. Although not a speech perception test, a commonly used measure during the 1980's was the Sound Effects Recognition Test (SERT). The task required that the children match the presented environmental sound with the correct picture.

Closed-Set Tests

Numerous closed-set word tests exist and are included as part of the assessment battery pre- and post-implantation. The tests serve a dual function: they provide a baseline by which post-implantation improvement can be documented over time and they furnish audiologists and speech patholo-

gists with information helpful for device programming and rehabilitation. Cumulatively, the tests assess prosodic features, speech features, word identification and sentence recognition and numerous tests need to be administered in order to obtain a complete profile of a child's auditory capabilities. The Early Speech Perception Test (ESP) (Moog and Geers, 1990) has two versions: The Standard version and the Low-Verbal version. The Standard ESP is designed for hearing impaired children six years of age and older while the Low-Verbal form is for younger children who do not have the linguistic competence to perform the Standard test. The test consists of sub-tests which assess pattern perception, spondee identification and monosyllabic word recognition. Children are required to choose the correct response from four picture response cards (small toys in the Low-Verbal version) out of a possible twelve, with a chance score of 25%. Based on the results, the children are placed in categories of speech perception skills ranging from no pattern perception to consistent word identification.

The Indiana Minimal Pairs test (Robbins et al, 1988) requires that the child point to the picture of the spoken stimulus. The test consists of twenty pairs of words with one phonetic feature differing in a pair: consonant manner, voice or place and vowel height or place. The test allows the examiner to determine if a child can detect a single difference between two words. It is essentially a feature perception test utilizing a closed-set format.

The Northwestern University Children's Perception of Speech test (NU-CHIPS) (Elliot and Katz, 1980) measures word recognition in stimuli using the most frequently occurring phonemes in the English language. The subject is required to choose the correct response from a group of four pictures giving a chance score of 25%. The Word Intelligibility by Picture Identification (WIPI) (Ross and Lerman, 1979) is a six-choice picture test designed for children above the age of five. The test was originally standardized on moderately hearing impaired children but can be used with profoundly deaf children who are familiar with the vocabulary.

The Imitative Speech Pattern Contrast (IMSPAC) (Boothroyd, 1991) test is a pediatric version of the SPAC, a four-alternative-forced-choice test designed to assess segmental and suprasegmental contrasts. In the pediatric version, the child is asked to repeat stimuli. The responses are recorded and listeners are asked to choose the correct production from four possible choices

which vary by a single phoneme. The test, conceptually designed to eliminate linguistic content, was too time-consuming and listener dependent to enjoy widespread usage. A three-interval-forced-choice speech pattern contrast perception test (THRIFT) (Boothroyd, 1986) was also devised to obtain speech feature information from children. This test is similar to the SPAC in theory but with a different task: the child hears three stimuli and must identify the one that is different from the other two. Again, the length of the test and the lack of the ability of children to consistently perform the task, precluded widespread usage. The applicability of a computerized version of the THRIFT geared to a wider age spectrum is currently undergoing field trials at several implant centers.

The Ling five-sound test (Ling, 1989) has been described as an informal and quick screening method of assessing a child's ability to detect differences between phonemes. The five phonemes typically used are 'a, e, oo, sh, s' although many clinicians add other phonemes to expand the limits of the frequency range of the acoustic properties. At the initial stimulation of a cochlear implant, a child might be asked to simply detect if he hears a given phoneme. As time progresses, however, the child might be asked to repeat the individual phonemes which upgrades the task from the simpler detection level to the level of identification.

Other closed set tests do exist but have been utilized infrequently with cochlear implant candidates and recipients. Information regarding these additional measures can be found in the reference text listed at the end of this monograph.

Although not a test of prosodic features or closed-set speech perception, the Meaningful Auditory Integration Scale (MAIS) (Robbins et al, 1991) and the Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS) (Zimmerman-Phillips et al, 1998) are parent questionnaires which were developed to assist in determining whether a child is maximizing use of an implant or any other type of sensory aid. The MAIS attempts to assess the auditory skills of a child based on the daily observations of the parents. The test consists of ten questions which probe the child's auditory responses under a variety of common daily situations. Since the results have been shown to correlate with results obtained on formalized tests of speech perception in children using cochlear implants, the test has been used in pediatric clinical trial protocols for several cochlear prostheses. The

IT version is an adaptation of the test with questions geared to children under two years of age.

Open-Set Tests

Table 2 lists some of the more recent and commonly used pediatric open-set tests. Open-set tests are divided into word and sentence recognition measures. The word recognition tests include the Phonetically Balanced Kindergarten Test (PBK) (Haskins, 1949), the Multisyllabic Lexical Neighborhood Test (MLNT) (Kirk et al, 1995) and the Lexical Neighborhood Test (LNT) (Kirk et al, 1995). The PBK word test consists of four lists of 50 monosyllabic words each. The words are presented at suprathreshold levels and scoring is in percent word and/or phonemes correctly identified. The MLNT contains a 15-item 'easy' list (Level 1) and a 15-item hard list (Level 3). Both lists consist of two- and three-syllable words. Level 3 is given when a score of 20% is achieved on Level 1. The LNT is a similar design but is a monosyllabic word test consisting of two 50-word 'easy' lists (Level 1) and two 50-word 'hard' lists. As with the MLNT, Level 3 is administered when a score of 20% is achieved on Level 1.

Open-set sentence recognition tests include the GASP (Erber, 1982), the Common Phrases Test (Osberger et al, 1991), the Bamford-Kowal-Bench Sentences (BKB) (Bench et al, 1979) and the Hearing In Noise Test for Children (HINT-C) (Nilsson et al, 1994). The GASP sentences consist

Table 2. Frequently used open-set speech perception tests for children

Test	Reference
Bamford-Kowal-Bench Sentences	Bench, Kowal and Bamford (1979)
Common Phrases Test	Robbins, Renshaw and Osberger (1988)
Glendonald Auditory Screening Procedure	Erber (1982)
HINT-C Sentence Test	Nilsson, Soli and Sullivan (1994)
Lexical Neighborhood Test	Kirk, Pisoni and Osberger (1995)
Multisyllabic Lexical Neighborhood Test	Kirk, Pisoni and Osberger (1995)
Phonetically Balanced Kindergarten Word List	Haskins (1949)

of 10 questions such as 'what is your name?' and the child is required to answer the question accurately in order for the sentence to be scored as correct. The Common Phrases Test consists of six lists of ten questions/statements per list. The child is required to repeat the sentence and scoring is based on the number of sentences and/or words correctly identified. The original BKB sentence test was standardized in England on 8 to 16 year old hearing impaired children. Since some of the vocabulary was peculiar to children in the UK, several adaptations have become available for use in the United States. The test consists of 100 statements and the score is the number of key words correctly identified. The HINT-C was developed to provide a measure of speech reception for sentence material in quiet and in noise. The stimuli consist of 25 equivalent lists of 10 sentences each with a broad-spectrum noise. The noise is presented at a fixed level while the level of the sentences is varied according to the response of the child.

The above is only a partial list of the available phoneme, word and sentence tests designed to assess speech perception in the hearing impaired child. Despite the rather extensive appearing list, several problems related to the test battery still remain. First, the majority of measures described are not appropriate for very young children. For example, the GASP test was given to a group of 7 to 13 year old children. The HINT-C test was designed for children 6 to 12 years of age and the fact that the LNT and MLNT are based on the vocabulary of 3 to 5 year old normal hearing children, does not guarantee that deaf children have the necessary linguistic competence or cognitive skills to either understand the task or perform the test. The fact that the age boundaries for the implantation of children has decreased to 18 months of age with implantations being performed on children as young as 12 months old, underscores the dilemmas related to assessment: currently no realistic protocol exists which can assess the auditory perception of very young children as related to their ability to either obtain significant benefit from conventional amplification preoperatively or with a cochlear implant for the first few years postoperatively. Once the degree of hearing loss has been established by either conventional audiometric procedures and/or electrophysiologic studies, the decision to implant a very young deaf child is based on the assumption that a child with a severe-to profound hearing loss who receives a cochlear implant will have a better opportunity to develop auditory and linguistic skills than would a

child using hearing aids. This conclusion is based on published data comparing pre- and post-operative performance in children which we will review later on in this monograph.

SURGERY

Prior to the initiation of clinical trials with multichannel cochlear implants in children, several issues specific to the surgical procedure in the pediatric population were addressed. In 1986, a multidisciplinary colloquium was held to examine all aspects of cochlear implantation in children. At that time the pediatric protocol for the 3M/House single channel device, the only implant available to children in the United States, allowed for implantation of children no younger than two years of age. Despite the fact that the cochlea is adult size at birth, the recommendation of the surgical group was not to proceed with implantation in children less than two years of age. The surgical concerns were related to the possible difficulties which could potentially arise from skull maturation. O'Donoghue et al (1986) found that the distance between the placement of the electrode array and the receiver/stimulator increases approximately 1.5 centimeters from birth to adulthood with about half of the growth occurring during the first two years of life. The concern was that this growth in distance would tug on the electrode array causing it to either displace or extrude thereby affecting performance and requiring reimplantation. Recently, Roland et al (1998) conducted a study to evaluate the stability of the electrode array in children. Serial radiographic studies were obtained on young children who had received cochlear implants. The length of implant usage ranged from one to five years. Results show no movement or displacement of the electrode array over time thereby reducing the anxiety associated with the implantation of the young child. It is important to note, however, that the surgical procedure incorporated the placement of the receiver-stimulator in a bony well drilled to size and ties used to secure its position. In addition, there have been no reports of electrode extrusion from surgeons who have been implanting very young children.

The surgical technique itself does not significantly differ from that used in either older children or adults. Waltzman and Cohen (1998) reported on a group of children who were implanted below the age of two and found considerable overlap on several physical dimensions: the heads

of the younger children were often as large or larger than those found in two year old children and no difference was found in the facial recess. The surgical procedure for all devices is basically the same except where the physical characteristic dimensions of a device dictate modifications. In general in children, Cohen (1998) recommends a vertical postauricular incision and a well drilled down to dura. As with all young children with small heads, the implanted electronics package is basically placed vertically with the inferior portion sufficiently behind the auricle to allow for the use of the either ear level processors or the microphone case of the Nucleus devices. The exposure of the middle ear, the cochleostomy and the electrode insertion were no different than for the older child. Following insertion of the electrode array, the receiver/stimulator is placed in the well and tied down with nonabsorbable sutures and the flap is closed. An intraoperative x-ray is recommended to insure proper electrode placement. For a complete description of the surgical phase used in children, please see Cohen (1998).

Due to implant design differences and insertion techniques, it is suggested that surgeons who have no experience or opportunity to practice with a given device, attend a manufacturer sponsored surgical course to familiarize themselves with the procedures necessary for successful insertions.

DEVICE PROGRAMMING

Initial device stimulation occurs approximately one month following surgery. Since all current prostheses transmit via the use of magnets, it is desirable to allow sufficient time for the swelling around the surgical site to diminish as much as possible. This allows for most efficient coupling between the two magnets which, in turn, enables the clinician to obtain more accurate measurements. Although device programming is often one of the least discussed and emphasized portion of the implant process, it is most certainly one of the more crucial ongoing contributing factors to ultimate outcome. Although a number of different encoding strategies exist both within and between devices, the fundamental components involved in programming are the same: the establishment of an electrical threshold and comfort level for each electrode in the array. Currently, the Nucleus CI24M, the Clarion and the MedEl devices utilize a monopolar stimulation mode. The CI24M is also capable of stimulating in a bipolar fashion.

Ordinarily, a method of limits is used to bracket

the threshold which is defined as the lowest level that the child responds to the electrical stimulus 100% of the time. Maximum comfort level is defined as the maximum level of loudness that can be tolerated without discomfort on a given electrode. These two measurements determine the dynamic range for a particular electrode and are the measurements used by the computer to generate the program used by the child. Doubtless, if the thresholds and comfort levels, which are the basis of the program in the speech processor, are inaccurate, the quality of the sound delivered to the child will be amiss, as well. With an older child, the establishment of accurate thresholds is relatively uncomplicated since, like adults, they have extensive experience with routine hearing tests. Young children are often more challenging; however, it has been our experience that even very young deaf children can be conditioned to respond to low level sounds and are often able to do so with relative ease. As with routine hearing testing in the pediatric population, it is important to remember that responses are often individual to a given child. One child might blink, another might laugh or cry, another might open his/her eyes widely and so on. It is very efficient to have two audiologists participate in the programming sessions: one operates the computer while the second does play audiometry with the child and observes the responses.

Comfort or maximum acceptable loudness levels are then set for each electrode. Again, older children can often let the audiologist know when the sound is not too loud but the task is often more difficult with younger children. Initially, underestimating the comfort levels is preferable to frightening the child with a strange loud sound. Softer stimuli at first, with a gradual increase in the loudness of the signals, although frustrating for the parents, is often the most effective way to have the child adjust to the implant. A sweep of the comfort levels should be performed whenever possible to insure that there are no unfavorable responses to a given electrode.

It is usual that adjacent electrodes have thresholds and comfort levels that are similar or close. Thresholds that differ significantly from a neighboring electrode can alter the quality of the sound and can signal problems with a given electrode. It is essential that parents and professionals alike remember that the thresholds, comfort levels and dynamic ranges which are set during the early stages of stimulation are subject to change. The adjustments occur not solely because of the in-

creased reliability of the child's responses: the acceptance of louder sounds increases with length of usage and adjustment to the electrical signals. Additionally, there appears to be physiologic changes which do sometimes effect changes in thresholds over time. And the setting of thresholds and comfort levels is not the whole story. These psychophysical measures are used by the computer to create a program. Once the program is created and the child is presented with speech live voice, there may be adjustments to the loudness, etc., which are necessary to optimize the listening condition. Depending on the chosen processing strategy, there are numerous manipulations that can be made after the psychophysical measures are obtained and the program generated. Alterations to the SPEAK (spectral peak) coding strategy include a percentage increase in all threshold and/or comfort levels, modification of the frequency allocation to the electrodes, and a widening or narrowing of the pulse width. The possibilities with the CIS strategy in the CI24M device are more extensive: initial threshold and comfort level measurements can be made at slower and faster pulse rates to determine the most optimum initial setting. This requires loudness balancing across electrodes at both settings. Since the CIS strategy uses 6 to 8 electrodes, one has to determine which electrodes across the array will be allocated to specific channels. The shape of the gain and frequency response, the order of stimulation (base-to-apex, apex-to-base) and the jitter percentage can be manipulated with older children and adults. For instance, if a patient complains that the sound is too high in pitch, the stimulation rate can be reduced. Alternately, the addition of jitter can help reduce the complaint of 'buzzing' in slower pulse rates.

Parameter setting with the ACE (advanced combination encoding) strategy is similar. With adults, the first part of the procedure is to establish a preferred pulse rate remembering that a lower pulse rate allows the usage of a greater number of maxima: the higher the pulse rate, the lower the number of possible maxima. In addition to the manipulations described for the CIS strategy, optimizing the ACE map includes the deactivation of basal electrodes. With the Clarion device, parameters including electrode firing order, input dynamic range and frequency shaping can also be controlled to effect optimum results. Default settings for all parameters for each of the processing strategies have been established by both Advanced Bionics and Cochlear Corporation and until such time as the child or parent can

provide some form of reliable feedback, it is advisable to use these default settings when programming young children.

In addition to standard behavioral techniques, several objective measures of programming have been attempted with very young children who are unable to respond subjectively. The electrophysiologic measures include electrical auditory brainstem responses (EABR), electrical stapedius reflex measures (ESR) and, most recently, neural response telemetry (NRT).

Shallop et al (1991) examined the relationship between EABR thresholds obtained intraoperatively and threshold and comfort levels obtained during device programming. He found that the EABR thresholds were more closely aligned with behavioral comfort levels and frequently exceeded the comfort levels by more than 20 units. Similarly, Mason et al (1993) found that intraoperative EABR levels exceeded programmed measured threshold units by an average of 35 units in 24 children from 2 to 11 years of age. Brown and colleagues (1994) found results in adults and children which were similar to those of Mason and colleagues (1993) in that EABR thresholds invariably fell above the objectively programmed thresholds but below the comfort levels; that is, within the dynamic range. To summarize, although EABR measures might provide some useful information regarding starting points for programming young children and the difficult to test population, they cannot be used to calculate exact threshold and/or comfort levels in device setting.

The predictability of ESR for behavioral threshold and comfort levels is likewise questionable. Several investigators (Jerger et al, 1986; Battmer et al, 1990; Hodges et al, 1997) have shown close correlations between ESR and comfort levels. Spivak and Chute (1994), however, found that the parallel between ESR and comfort levels can vary considerable across patients with only 50% of their subjects showing agreement between ESR and behavioral comfort levels.

Neural Response Telemetry (NRT) offers some promise for programming application. The Nucleus CI24M cochlear implant is equipped with a bi-directional neural response telemetry system which permits measurement of electrical action potentials from within the cochlea. Using this technique, one can obtain information regarding the response of the auditory nerve to electrical stimulation and the integrity of the prosthesis. This procedure allows adjacent electrodes to record the voltage in a stimulated electrode pair following

the presentation of the stimulus. Various parameters of the amplified and averaged waveforms, consisting of N1 and P2 peaks, can then be analyzed and evaluated. For example, as the stimulus level is increased, the amplitude of the EAP also increases allowing for the study of growth and recovery functions. This is a particularly attractive alternative since it does not require any additional external equipment and can be performed in an awake patient with no discomfort. The use of NRT as a possible programming tool is just beginning to be examined. In 1994, Brown and colleagues found a correlation between behavioral thresholds and EAP thresholds. Hughes et al (1998), measuring the EAP in children with the CI24M, found that the EAP thresholds consistently settled between the behavioral threshold and comfort levels i.e. within the dynamic range.

Although electrophysiologic methods, particularly NRT due to its ease of use, have the potential to assist in programming young and/or uncooperative children, the lack of definitive correlations between behavioral and electrophysiologic thresholds and comfort levels limits the applicability. It is expected that as more data is collected related to NRT, the ability to use it as a part of an effective programming battery will be enhanced.

Since valid electrical thresholds are a precursor to accurate sound access, and since research has shown that psychophysical threshold measurement in children change over time (Shapiro and Waltzman, 1995) it is advisable to schedule follow-up programming visits on a consistent basis. A basic post-initial stimulation timetable can be as follows: 2 weeks, 4 to 5 weeks, 3 months, 4.5 months, 6 months, twice between 6 months and 12 months and at 12 months. Additional visits are advisable when a parent or therapist reports some negative change in either the child's perception or production behavior including: reduction in auditory responsiveness and/or understanding, rise in need for repetition, poorer vocal quality, prolongation of vowels and omission or addition of phonemes or syllables. It is important to remember, however, that following a change in the program, a child may experience a temporary decrease in auditory and production skills. These decrements in competence will disappear once the child has adjusted to the new program: it is therefore critical to appreciate the patterns and needs of each child when a programming schedule is prepared since adjustment periods do differ between children. It is also important to realize that not all changes in auditory, speech and linguistic behaviors are bound

to, or correlate with, specific programs. As an example, it is usually unwise to attempt to manipulate specific electrode parameters in an attempt to 'fix' specific phonemes if the changes upset the balance of the entire program. Wish it were that fine-tuning of a program alone could account for ultimate performance! Input from parents and therapists is invaluable in guiding the audiologist regarding programming needs. Parents of virtually every hearing impaired child have spent a lifetime advocating and fighting to have the educational, social and personal needs of the child met; however, to serve the best interests of the child, there are principles of programming, including accurate psychophysical measures and loudness balancing, that the audiologist needs to adhere to in order to provide the child with the best possible access to sound. Ultimately, it is most beneficial to the child if the audiologist determines what type of program change is necessary based on a combination of objective and subjective variables.

EDUCATION AND TRAINING

The child has been evaluated, deemed an appropriate candidate, implanted and programmed. So . . . what happens next? Well, what transpires following the initial stimulation and for years to come, determines to a significant degree the nature and type of communicative ability that a child will develop: all things being equal, the communication mode and educational path which ensue have a marked effect on both the speech perception and production skills that emerge following implantation.

The effects of training and education began to surface in the early days of implantation of children. Quittner and Steck (1991) examined eighteen 3M/House and eleven Nucleus users who were prelingually deafened and had a mean age at time of implantation of 9.3 years (range = 5 to 16 years). They found that the children who were oral communicators (OC) were better users of sound than the children who used total communication (TC). Staller et al (1991a) reviewed the results from 80 children who participated in the original pediatric clinical trials for the Nucleus device. Fifty five percent were prelingually deafened while 34% were congenitally deaf. The mean age at time of implantation was 9.10 years and 38% were OC, 43% were TC and the remaining 19% used cued speech as their primary means of communication. They found that the OC group had consistently higher speech perception scores than either of the

other two groups. More recently, Lusk et al (1997) performed a multivariate analysis on results from 85 children implanted with the Nucleus device and users for two years. Sixty-six were congenitally deafened and 19 had acquired deafness. The mean duration of deafness was 6.64 years and the mean age at time of implantation was 7.09 years including 25 children implanted prior to age five; 32 were implanted between 5 to 10 years of age and 21 were implanted above the age of 10. Three factors correlated with postoperative performance: duration of deafness, early implantation and oral education. Osberger and Fisher (1997) examined 44 prelingually deafened children who had been implanted with the Clarion multichannel cochlear implant: 25 (57%) were OC and 19 (43%) were TC. The mean age of onset of deafness was 6 months and the mean age at implantation was 5.6 years for the OC group and 5.7 years for the TC group. Evaluations at the 6-month and 12-month postoperative intervals revealed significantly higher scores for the OC group on the perception of monosyllabic words as measured by the PBK test. Similarly, Dowell et al (1997) found that OC children had higher scores on speech perception tests than did TC children. Meyer and colleagues (1998) examined the differences between OC and TC children on the Common Phrases test. When the stimuli were presented in the auditory-only condition, the OC children performed better than the TC children. Following one year of usage, the OC children scored an average of 40% whereas the TC group averaged 35% following four years of usage.

In addition to speech perception skills, several investigators have begun to explore the effects of education and communication mode on speech production and language development. In 1994, Osberger and colleagues found that OC children showed greater improvement in speech production skills that did TC children. More recently, Svirsky et al (1998) found that speech intelligibility correlated with open set speech perception for both OC and TC groups. If the PBK phoneme score was below 40%, no difference in intelligibility existed between the groups; however, OC children with PBK phoneme scores above 40% had more intelligible speech than TC children with the same scores. They also found that language development was highly correlated with open-set speech perception in OC but not in TC. This makes sense since the better an OC child can hear, the more likely they are to develop sound linguistic skills; undoubtedly, the TC children are able to develop comparable language skills via the manual mode.

Archbold (personal communication) examined the three year postoperative performance of implanted children and found that the OC children outperformed the TC children on measurements of both speech perception and production. An additional interesting finding in this study was that there was no significant difference between the progress of children who had been in oral schools prior to implantation and remained there versus children who had been in TC settings prior to implantation but switched to OC settings at some point postoperatively: both groups did better than children who were still in total communication sites at the three year postoperative interval.

Undeniably, there are many confounding issues: device, processing strategy, age at implantation, length of deafness, medical issues, family commitment, etc. But it is important that parents be made aware of these outcomes during counseling prior to implantation. Although all children, no matter what the chosen mode of communication, will show improved communication skills following implantation, parental expectations should be directed, at least in part, by the selected communication path. Research outcomes could potentially be very helpful to parents as they embark on the decision making process regarding the direction of their children's education and training.

SPEECH PERCEPTION RESULTS

The authors will not attempt to review the vast amount of literature which has been published on the perception of sound by children using cochlear implants. Much of the early published results focused on the perception of the suprasegmental portions of the speech signal, closed-set perception and visual enhancement. The advancement of speech processing strategies has shifted the focus of recent investigations to open-set perception of phonemes, words and sentences. The authors will briefly review some of the relevant past work but concentrate on the more recent outcomes related to the development of open-set speech recognition skills in the pediatric population.

The ability to detect rhythmic, stress and intensity patterns of speech is the simplest form of perception. Although these time/intensity cues may help with the identification of a speaker, the ability to distinguish between a question and statement and assist with lipreading tasks among other things, they cannot facilitate the understanding of open-set speech stimuli. As discussed in the Assessment section of this manuscript, several tests

have been designed to examine perception of su-
prasegmental aspects of speech. The Monosyllable-
Trochee-Spondee test, the Change/No Change and
some subtests of the Minimal Auditory Capabili-
ties (MAC) battery (intended for adults but can
be used with children) have been the most com-
monly used tests. The MAC battery measures in-
clude the number of syllables and the male/female
tests. During the initial pediatric clinical trial for
the Nucleus device, the MTS test was a part of
the protocol and Staller et al (1991b) reported on
the results of 83 children who participated in the
clinical trials. Following one year of usage, the mean
percent correct score was 64%. The highest scores
were obtained by postlingually deafened children
although many children in the postlingual, prelin-
gual and congenital groups scored at chance level.
Osberger et al (1991) described the results on sev-
eral of the suprasegmental tests for 28 children
implanted with the Nucleus device. On the Change/
No Change test (chance = 50%), the scores aver-
aged 89% for syllable length, 87% for vowel
height, 85% for fundamental frequency, 82% for
gender, 82% for vowel place, 80% for consonant
manner and 64% for intonation. When Osberger
et al (1991) compared a smaller group of pediatric
Nucleus users to 3M/House implantees, she found
that the Nucleus users scored significantly better
on both the MTS and Change/No Change tests
confirming better performance with multi-channel
cochlear implants. Although these data provided
interesting insights, they were collected after only
short-term experience with implant usage and did
not shed much light on the ability of the implant
to access information which would account for
speech understanding. The goals for pediatric co-
chlear implantation at that time were modest:
those involved were not at all certain that the pro-
cessing capabilities would allow for open-set
speech recognition in postlingually deafened chil-
dren or that the devices would be capable of al-
lowing a young child to develop oral linguistic skills.
As time progressed, it was evident both from the
evolution of processing and encoding capabilities
of commercially available implants to results ob-
tained on adults and initial outcomes on children,
that expectations needed to increase and assess-
ments needed to encompass a broader range, and
higher level, of skills. Word and sentence recogni-
tion using closed set measures with restricted re-
sponses are not an indication of function in unre-
stricted listening situations. Hence the need for
open-set tests which can more accurately estimate
speech perception in everyday listening situations.

Open-Set Speech Recognition

The focus of cochlear implantation in children
was rapidly evolving, encompassing many areas
which were shifting simultaneously. Several inves-
tigators (Staller et al, 1991a, b; Fryauf-Bertschy et
al, 1992; Waltzman et al, 1992, 1994; Miyamoto
et al, 1993; Gantz et al, 1994) began assessing post-
operative open-set discrimination in pediatric users
while, at the same time, examining the effects of
implantation on the congenital and prelingually
deaf population implanted at a variety of ages.
Each of the studies showed significant improve-
ment in all areas of speech perception including
prosodic features of speech, vowel and consonant
perception and, particularly, open-set phoneme,
word and sentence recognition.

Staller et al (1991a, b) studied a group of chil-
dren who were part of the initial Nucleus pediat-
ric clinical trials. They identified duration of deaf-
ness as a significant contributor to postoperative
open-set speech recognition in children whose
mean age at time of implantation was 9.8 years;
that is, a child with a shorter duration of deafness
was likely to have better postoperative perfor-
mance. In the Miyamoto study (1993) the average
age at implantation was six years with a mean du-
ration of deafness of five years. In the Waltzman
et al study (1994), the mean age at implantation
was two years and the mean duration of deafness
in the non-congenital prelingual group of children
was one and a half years perhaps accounting, in
part, for the better auditory-only open-set perfor-
mance of the children in the Waltzman et al study.
Both Gantz et al (1994) and Waltzman et al (1994)
showed significant and non-plateauing open-set
auditory skills. On the PBK monosyllabic word
test, Gantz et al found that four years postimplan-
tation, 80% of the pediatric subjects had measur-
able scores ranging from a mean of 5% for the
prelingual group to 10% for the congenital group.
In the Waltzman et al (1994) study, following
three years of usage, the average word score on
the PBK test was 47%.

In 1997, in response to the need to further de-
lineate and separate variables for analysis, Waltz-
man and colleagues reported on the development
of auditory-only open-set skills in congenitally
deaf children implanted below 5 years of age. Re-
sults over a five year postoperative period indi-
cated a continuous and steady growth of open-set
speech recognition as measured on the PBK test:
the average word scores for each of the years
were 5%, 16%, 44%, 33% and 58%, respectively.

It is important to note that 37 of the 38 children in this study were oral communicators—only one child used total communication. In an extension of the study, Waltzman and Cohen (1998) assessed the open-set recognition capabilities of nine children implanted below the age of two. Following at least two years of implant usage, the monosyllabic word scores ranged from 40–80%. Since the children were all evaluated, implanted, programmed and followed at the same institution and received the same type of training, the results of this young group were compared to the results obtained in the 1997 study on children implanted between the ages of 2 to 5 years. Although the differences in the number of subjects prevented statistical analysis, it appears that the younger children achieve high levels of open-set perception at a younger age that the 2–5 year old group of children. It is likely, however, that over time the two groups of children will achieve similar levels of auditory perception. In any event, the scores increased for all children with length of usage. Similarly, Fryauf-Bertschy et al (1997) found a significant difference in performance on PBK words following three and four years of device usage between children implanted above the age of 5 years and those implanted younger than 5 years of age. Individual scores for the younger group reached approximately 70% whereas the highest score achieved for the older group was approximately 40%.

Similar, and sometimes enhanced, auditory development in children is being reported with the use of advanced encoding strategies. Preliminary data were reported by Cohen et al (1999) on children above the age of 5 years who were implanted with the Nucleus CI24M device. Preoperatively, 3 of 7 children scored 8% or more on the PBK word test. Following only six months of device usage, six of the seven children achieved scores above 8%. Additionally, Waltzman and Cohen (1999) reported on the acquisition of open-set auditory skills in long-term deafened children using the CIS strategy as implemented in the Clarion device. Preliminary three-month post-implant data reveal that 15 of 21 children with a length of deafness of 10 years or more, had some amount of open-set speech recognition. Ten of the 15 subjects are OC and had the highest perception scores. Similar trends are beginning to emerge with the Nucleus CI24M. With the focus being on the implantation of young children, it is encouraging to observe that the more advanced strategies offer the long-term deafened population, which previously had a poorer prognosis (Fryauf-Bertschy

et al, 1997) the possibility of improved auditory perception.

In summary, the development of auditory-only open-set phoneme, word and sentence perception skills in congenital, prelingual and postlingual pediatric cochlear implant users has been well documented. Similar, and sometimes enhanced, performance is being reported with short-term and long-term deafened children with the commercial availability of updated processing strategies. What is equally evident, however, is the wide range of postoperative performance that has been described. It is only recently that factors, in addition to processing strategies and surviving neurons, have begun to be explored as variables affecting outcome. These variables include, but are not limited to, age at implantation, length of deafness, mode of communication, educational setting, training, programming techniques, surgical issues and family interactions. It is equally important to realize that as the design of the devices becomes increasingly more advanced, the role of a given variable might take on lesser or greater importance in the equation.

Although many questions related to factors which influence ultimate perception performance still remain, there is little doubt as to the efficacy and safety of cochlear implants in children. We now need to proceed to the next logical step—the children can 'hear' but does this hearing afford them the opportunity to develop oral language and speech production skills? Since language is the basis of communication, learning and achievement throughout life, it, by definition, becomes a critical missing component in the development of a profoundly deaf child.

LANGUAGE DEVELOPMENT AND SPEECH PRODUCTION

Receptive and expressive language development in the deaf child has been shown to be both delayed and aberrant in its development (Boothroyd et al, 1991; Geers and Moog, 1994). In fact, the average congenitally deaf child acquires language at one-half the pace of a normal hearing child (Robbins et al, 1997) and may never catch up in terms of the development of conceptual or abstract thinking which is based on a firm command of the English language. Limitations are also noted in their reading and writing skills which have been shown to be severely compromised. In fact, the average profoundly deaf high school graduate is capable of reading at the third grade level which severely

limits their professional opportunities (Schildroth and Karchmer, 1986).

Since much of the early research in pediatric cochlear implantation focused on the efficacy of the devices, very few early studies exist on the development of language skills post-implantation. The few documented analyses of postoperative performance showed an average increase in language learning which exceeded predicted amounts and surmounted levels achieved by hearing aid users (Geers and Moog, 1994).

Recently, investigators have concentrated more effort to document the language growth in pediatric implantees. Robbins et al (1997) compared the rate of language growth on the Reynell test in a group of profoundly deaf children implanted with a cochlear implant and those who did not receive implants. They found that after one year, the implanted group acquired language at a rate equal to normal hearing children, that is, twelve months of growth in a twelve month period of time, a rate far more rapid than what was expected of the non-implanted children who were predicted to gain six months of language in a twelve month period.

Brackett and Zara (1998) assessed the language development of 33 profoundly deaf children implanted at NYU Medical Center. The children were between the ages of 2 to 5 years at time of implantation, were followed for a period of three years and were oral communicators. Both receptive and expressive vocabulary were measured. The mean growth over the 36-month period was 33 months for the receptive portion and 48 months for expressive vocabulary. During this period of time, the children developed the ability to use simple sentences with appropriate verb tenses and higher level grammatical components were beginning to appear.

The above studies were performed on children who were implanted with the Nucleus device, and who, for the most part, were programmed using a multipeak coding strategy. Robbins et al (1998) assessed a group of children implanted at a mean age of 38 months with the Clarion cochlear implant and programmed using the CIS coding strategy. Following six months of device usage, the average rate of receptive and expressive language growth was nine months, exceeding the rate at which normal hearing children learn language.

Robbins et al (in press) summarized the prevailing language development studies as follows:

1. Improved speech processing strategies provide more language enhancement,

2. Children with cochlear implants outperform their profoundly deaf peers who use hearing aids,

3. Cochlear implants allow deaf children to begin to learn language at a rate equal to that of normal hearing children,

4. Initially, many implanted children remain delayed in their language skills even after implantation,

5. A wide range of language benefit is observed across children,

6. Language skills improve in both OC and TC implanted children, though the oral language skills are consistently better in the OC group,

7. Younger age of implantation provides a better chance for language development.

The capacity to be understood when speaking is central to the ability of the deaf child to function independently in a normal-hearing educational and social environment. In 1994, Osberger and colleagues at Indiana University studied the intelligibility of profoundly deaf implanted children over a five-year period and compared these results with those of hearing aid users. The hearing aid users were divided into three groups: the 'Gold' users had unaided pure tone averages of 90–100 dBHL, the 'Silver' users had unaided pure tone averages of 100–110 dBHL and the 'Bronze' users had unaided thresholds above 110 dBHL. At each test session, the children were required to repeat ten sentences. The recorded responses were listened to by a group of unsophisticated listeners and analyzed as percent words correct. At 4 years post-implantation, the mean intelligibility was 40%—about 20% above the scores for the Silver hearing aid users but below the intelligibility ratings for the Gold hearing aid users. Similar results have been reported in later studies conducted at Indiana University (Miyamoto et al, 1996; Miyamoto et al, 1997). Svirsky (personal communication) examined speech intelligibility of Nucleus MPEAK users and found some correlations between perception and intelligibility. The children who scored 40% or better on PBK phonemes and were orally trained had more intelligible speech than children with poorer PBK scores or than children with equal scores who were TC users.

Brackett and Zara (1998) used the CID Phonetic Inventory Sample to assess production of 33 children implanted between 2 to 5 years of age at NYU Medical Center. The mean preoperative vowel production score was 24%; three years postoperatively, the children produced 88% of the

vowels. Similar gains were noted for consonant production: the average preoperative score was 8% and the average three year score was 69%.

As with the language studies, the data reported reflect results with 'older' speech processing strategies: predominantly the Nucleus MPEAK. In addition, many of the children were implanted when they were older. In 1998, Svirsky and his colleagues reported on the intelligibility of children implanted prior to 6 years of age and programmed with either the SPEAK strategy in the Nucleus device or the CIS strategy as implemented in the Clarion device. As would be anticipated, after several years of implant use, these children were more intelligible than previously reported: their intelligibility was similar to that of Gold hearing aid users.

The global improvements in perception, language development and production are certainly encouraging. Undoubtedly, the improvements in the design and processing capabilities of the implants and the earlier ages of implantation are two pivotal elements in this evolution.

PARENTAL INVOLVEMENT AND COUNSELING

The nature and strength of the commitment of parents, siblings and extended family of the child under consideration for implantation is central to the success of the process. The parents who are in the position of deciding whether or not a child is to be implanted should have access to all information which could potentially assist them with the decision-making process. There are many issues which need to be addressed including, but not limited to, the following:

1. Alternative treatment for the child including the use of conventional amplification or choosing a non-auditory path. Parents should be informed regarding the use of manual communication as either the sole means of communication or as an adjunct to the cochlear implant.
2. An explanation of the entire 'process' of implantation from beginning to end.
3. A complete description of the implant devices available to the child and the differences between them.
4. Based on published or unpublished data, a realistic assessment of the possible perception, language development and speech production benefits to the child post-implantation versus

those that can be obtained with a hearing aid. The discussion and prognosis should be tailored to the specific child wherever possible in terms of degree of hearing loss and perceptual skills with hearing aids, age at implantation, length of deafness, type of communication mode, educational placement, rehabilitation, etc. It is of vital importance, however, that all concerned be aware of the lack of ability to predict outcome with any certainty. Very often parents would like guarantees regarding the timetable for development of specific perceptual and linguistic skills: it is unwise under any circumstance to predict when, or if, specific skills will emerge. There are simply too many as yet undefined influences which no doubt affect performance to an unknown degree (see Table 3). When implanting a very young child, it is often impossible to diagnose learning disabilities and other neurological, cognitive, etc. which are possible impediments to optimal postoperative performance. The preoperative decision making period is the best time to discuss these important issues as it may help to avert disappointment or unexpected concerns later on.

5. A full description of the surgical procedure including all potential risks. Post-implant activity limitations should also be discussed since they are often individual to a given surgeon. Many surgeons will place few or no restrictions on activity whereas others might recommend avoiding activities such as wrestling, soccer, etc. In any event, helmets should be recommended during bicycle riding, baseball, etc.

The chance of device failure should be included in the discussion. Although the percent of failures of the internal system is very small with the current commercially available, they can, and do, occur. Parents need to be made aware of this possibility and know how the situation will be handled by the implant team should a failure occur. Of course, external equipment breakdowns and cord replacements, etc. should be addressed and the possible costs incurred should be summarized.
6. The nature, importance and frequency of programming sessions.
7. The educational options available to the child and the accompanying consequences. It is important for the parents to understand

Table 3. Variables affecting performance in children

1. Implant technology
2. Neuronal survival
3. Sensory deprivation
4. Plasticity of auditory system
5. Length of deafness
6. Age at implantation
7. Etiology of deafness
8. Criteria for implantation
9. Preoperative hearing levels, speech perception and linguistic abilities
10. Measures and techniques used to assess performance
11. Multiple handicaps
12. Surgical issues
13. Device programming
14. Device malfunction/failure
15. Mode of communication
16. Type/frequency of training
17. Education setting
18. Parental/family expectations, motivation, etc.
19. Consistency of follow-up.

that the choice of a particular school is not irreversible-depending on the progress of the child, the educational setting can be changed.

8. Decisions regarding, and arrangements for, post-operative rehabilitation should be finalized prior to surgery, whenever possible.

9. Prognosis and expectations.

It is particularly vital that the parents comprehend the time commitment and the labor-intensive nature of the procedure—not only for the professionals but for the family, as well. There has to be an awareness that they are active participants in the rehabilitation process: working with the child does not begin and end either at the implant center or during rehabilitation sessions. The way in which the family speaks and relates to the child contributes considerably to the eventual outcome. It is best to discuss all of these issues with the parents prior to the final decision regarding implantation so that the choice is indeed based on informed consent.

CONCLUSION

It is our desire that the reader come away with some basic concepts:

1. Cochlear implants provide children with access to sound not available through other sensory aids.
2. This access to sound provides the best opportunity for children to develop auditory perception, oral language and speech production to a degree sufficient to allow for independent functioning in a hearing world.
3. The degree to which the post-implant auditory and speech skills emerge is dependent on numerous internal and external factors, in addition to the implant itself.
4. The nature and scope of the field of cochlear implants is dynamic. The future, benefits and limitations are to a significant degree dependent on the evolution of device technology. It is conceivable that future technological developments will yield increased benefit to current and future implantees and be of value to a broader segment of the hearing impaired population and be less dependent on external variables.

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